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PIBIC Last Name - Mendieta First Name - Luana Middle -

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ELECTRORETINOGRAPHIC FINDINGS IN PATIENTS WITH USHER SYNDROME

L. Mendieta, A. Berezovsky, P. Sacai, J.M. Pereira, S.R. Salomão. Clinical Electrophysiology of Vision Lab, Dept. of Ophthalmology, Federal Univ of São Paulo, São Paulo, Brazil

Purpose: The Usher Syndrome (US) is a group of genetically and clinically distinct autossomal conditions, characterized by a sensorineural hearing loss accompanied by a retinal dystrophy indistinguishable from retinitis pigmentosa. There are two distinct forms of the disease: a common severe congenital form (type I) and a less common milder form (type II). The purpose of this study was to analyze full-field electroretinogram (ERG) responses and visual acuity scores between those two groups of patients with US. Methods: 16 patients previously diagnosed with US were referred for ERG testing (mean age at test = 25.1 ± 18.1 years) allocated in two groups. 12 patients with type I (mean age at test = 19.8 ± 15.4 years) and 4 with type II (mean age at test = 40.7 ± 19.1 years). Full-field ERGs were obtained in one eye through a dilated pupil after 30 minutes of dark adaptation according to ISCEV protocol. ERG and visual acuity outcomes were analyzed between group with US I and Group with US II by the t-test. Results: Scotopic rod and maximal (mixed cone and rod) responses were non-detectable both in US I and US II patients. The only detectable response was the 30Hz flicker photopic cone ERG, disclosing a severe reduction in amplitude and delayed implicit time. The T-test showed that those findings were comparable for the two groups. The mean visual acuity (VA) was comparable for the two groups, with US I showing 0,42 logMAR (20/50 Snellen equivalent) and US II showing mean VA of 0,45 logMAR (20/55 Snellen equivalent). Conclusions: Absence of rod function and severe cone ERG abnormalities were found in types I and II of Usher syndrome. Visual acuity was relatively preserved in these patients. However, night blindness seems to be an earlier finding in Usher type I.

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